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SPORADIC CRETINISM,

AND ITS DISTINCTION FROM FORMS OF IDIOCY AND OTHER DISEASES.

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The study of sporadic cretinism in this country received its impetus during the last few years from the writings of William Osler, who in 1893 published the first systematic analysis of the disease as we find it in America. The history of sporadic cretinism in America before this brochure appeared was confined to a few scattered cases published at intervals of years in the journals. Preceding the publication of the original three cases of Osler we find that A. Jacobi presented in his clinic on diseases of children a cretin eight years of age, with some data as to the history of the case. This is the first case published in America of cretinism emanating from a leading clinician. Moreover, it would seem that all statements preceding this publication of the occurrence of endemic or sporadic cretinism in America are unreliable. Immediately preceding the publication of Osler's cases we find two authentic cases

of sporadic cretinism recorded by Lloyd in the international clinics, and one case of congenital cretinism, published by Townsend, of Boston. Following the publication of Osler's paper, a number of scattered cases of sporadic cretinism appear in the recent literature. In addition to those published from other clinics in Osler's *résumé*, Rotch and Bullard, of Boston, Booker, of Baltimore, we have those of Northrup, Peterson, Noyes, Fruitnight, and Osgood Mason (1894). The author wishes also to state that in March, 1896, he presented two cases at the New York Academy of Medicine. If we turn to other countries we find that of late years the publication of cases of sporadic cretinism has increased also, and may be traced directly to the interest surrounding these cases as a direct result of the experimental work of Horsley and the clinical work of Reverdin, Simon, Kocher, and Ord. Within recent years we have cases published by Hillier (1893), Paterson (1893), Escherich (1894), Lebreton (1895), Sinkler Wood (1893), Ord (1893). Prior to 1871, we must be very cautious in accepting the publications on sporadic cretinism, for, while cases are published by Norris (1848) and Rees (1851), it would seem that the credit of clearly distinguishing the sporadic from the endemic forms of cretinism is awarded to C. Hilton Fagge (1871). This author not only published cases of sporadic cretinism, but sharply defined cases of cretinism with goître and those without goître, some of his cases being also idiotic. This it seems completes the historical *résumé* of sporadic cretinism. It will be seen that it is not so very extensive, though vast credit must be given to those who were the pioneer observers. Much confusion has arisen from the fact that sharply defined clinical pictures of sporadic cretinism

are absent in many publications where forms of idiocy, etc., have been included under the head of cretinism.

Definition.—Sporadic cretinism occurs away from the centres of endemic cretinism and in countries where, as in America, endemic cretinism is unknown. Though Hirsch includes America as among the countries afflicted with endemic goître and cretinism, Osler has taken much pains to analyze these statements and finds them unsupported by facts.

Sporadic cretinism, or infantile or congenital myxœdema, should now also include those congenital cases formerly reported as congenital rickets. Horsley insists upon the classification of these cases as forms of sporadic cretinism, as does also Barlow. Sporadic cretinism may therefore be congenital or fœtal, or appear some time after birth. In fact, the vast majority of the cases thus far published seemed to be healthy infants at birth, and from healthy parents, and within the first year the symptoms of sporadic cretinism or infantile myxœdema have appeared. Therefore, though the clinical pictures of endemic and sporadic cretinism are similar, they are not identical. Though sporadic and endemic cretinism may eventually be proved to be due to the same infectious agents (Kocher), yet endemic cretinism is now regarded as an advanced stage of a degeneration beginning with goître manifestations and resulting in the production of disturbances due to athyreosis, whereas sporadic cretinism occurs independently of goître, and, in fact, in the cases thus far recorded, the thyreoid has been atrophied or not in a marked way changed pathologically.

Endemic cretinism has occurred in all the sections of continental Europe. Its first mention we find by Forest or Felix Plater in the sixteenth century. In the latter

part of the eighteenth century Horace de Saussure, a traveler, described these unfortunates in the Alps. Following him, Ramond de Carbonier described the cretins of the Pyrenees. We then have the work of Fodère (1792), Wenzels, Joseph and Karl (1802), Iphofen (1804), Andrea (1814), Maffei Seizburg (1825), Hausler (1825), Wilke (1828), Troxler (1830–'36), Gross (1837), Demme (1840), Meyer (1845), Helferich (1847), Eulenberg and Marfels (1857).

It will thus be seen that cretinism has been endemic in Europe and has attracted attention as far back as the sixteenth century, but we find the first attempt to describe the disease from a pathological standpoint began with the work of Fodère, though Ackerman before him described the skull of the endemic cretin as a pronounced form of the rhachitic skull.

Fodère pointed out the connection between cretinism and goître, and insisted that cretinism was but an advanced stage of the goîtrous degeneration.

We must give to Virchow the great credit of having first studied the cretin skull. He showed that the peculiar form of the skull and expression of the endemic cretin physiognomy was due to a premature synthesis of the os basilare and the sphenoid, posticus and anticus. In this fact he seeks to find the principal cause of arrest of cerebral development in these unfortunates. Eulenberg and Marfels (1857) have confirmed these views of Virchow. Virchow describes the skull of the endemic cretin as for the most part of the brachycephalic type (short head). But we find endemic cretin skulls which include types of platycephalic skulls (flattened skulls), trochocephalic (round skull), and oxycephalic (pointed skull). In other words, the bony growth was the first element to engage

the attention of the early observers of the endemic cretins; the long bones were found shortened, plump, thickened, and otherwise deformed, without distinct ossification zones, with the heads of the bones flattened similar to the changes found in rhachitis (Klebs). As a result of the premature synthesis of the bones of the skull, we find in the endemic cretin the root of the nose broad, the lower jaw prominent (prognathous); and this latter Virchow and Grawitz think a characteristic of the physiognomy of the typical cretin. The low forehead, prominent cheek bones, with large alæ nasæ, give the whole face an appearance similar to the physiognomy of the Eskimo. In addition, we have the broad and thick lower jaw, the short and thickened neck, the flattened chest, and prominent abdomen. The arms and legs are short, the muscles flabby, and the gait wabbling. The skin shows the appearances described by Charcot as the *cachexie pachydermique*—an intense anæmia in which we have a greenish hue given to the skin. The skin is wrinkled and like in old persons, thickened, as if œdematous, and the hair is short, dry, and coarse; the genitals for the most part are rudimentary. Most cretins of the endemic types are deaf, and can not talk but in a few monosyllabic terms, mostly grunts, cries, or howls. If they do talk, they find most difficulty in pronouncing consonants. There is no aptitude for learning the simplest things, and no logical responsibility. In some cases there seems to be an everlasting torpor, in which the material being will sit for hours oblivious of surroundings. We find the thyreoid next engaged the attention of observers of this disease.

The Thyreoid Gland.—Fodère first insisted that endemic cretinism was an advanced stage in a series of de-

generations in which goître was the first manifestation. It was not until 1882 that J. Reverdin first noticed in extirpation of the thyreoid for goître the sequence of anæmia, œdema of the features, tumefaction of the eyelids, and feebleness of both physique and mind. The œdema and physical weakness much resemble those of Bright's disease. Kocher, of Bern, subsequently found, as also Wölfer, in looking over their cases of bilateral extirpation of the thyreoid, a condition exactly similar to that described by Reverdin; and Kocher gave this condition the name of cachexia strumipriva. It is a condition exactly identical with that found in the endemic cretin. The conclusion was at once inevitable that in those goîtrous subjects who were not the victims of cretinism there were still portions of the thyreoid gland which functionated and protected the individual from the cretinoid degeneration. Kocher gave to the peculiar state of the thyreoid gland which allowed the supervention of cretinism the term of "athyreosis." In his extirpations Kocher found that there supervened an anæmia of a progressive type, pains in the arms and legs, fatigue on the least exertion, reduction of body temperature giving rise to chilliness, slowness of speech, swelling of the hands and feet, with a peculiar dryness of the skin, and falling out of the hair. The anæmia progressed until the proportion of red to white cells was one in three hundred in eight months, and in one case the red blood-cells were reduced to 2,800,000. In individuals who were young, growth ceased. Kocher found that the thyreoid had a direct influence over growth of bone and blood formation.

Horsley, experimenting upon monkeys, found that he could produce a condition identical with cachexia strumi-

priva by extirpation of the thyreoid. This seemed to complete the connecting link between the functionating thyreoid and the condition known as endemic cretinism.

In looking back through the literature it was found that Sir W. W. Gull described a peculiar condition supervening in women, which he called the cretinoid state. One of his cases was a young girl, and one a woman of forty years. Ord redescribed a set of these cases and first gave to them the name of myxœdema. It was then seen that a form of cretinism could supervene in women and young people exactly similar to that of the cases of cachexia strumipriva independently of enlargement of the thyreoid, and these cases were concluded to belong to the same class as endemic cretins, the cretins operated on, or cachexia strumipriva, and due to the same cause—an athyreosis, an agent acting on the thyreoid, producing certain degenerations manifesting themselves either with a hypertrophy or atrophy of the thyreoid.

I have thus traced the early studies leading to the classification of sporadic cretinism among the cretinic or myxœdematous degenerations, and in 1887 the committee of the Clinical Society, London, classed infantile or congenital rickets or myxœdema, endemic cretinism and sporadic cretinism, under the same head as manifestations of the same influence acting on the functions of the thyreoid.

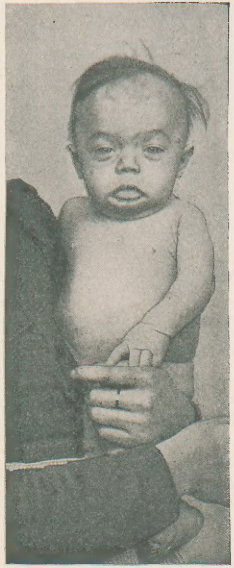
Sporadic Cretinism.—Hilton Fagge was the first to use the term sporadic cretinism, and by it we now understand a peculiar condition similar in many of its clinical features to endemic cretinism, supervening either *in utero* or some time after birth, and due probably to the same poison acting on the thyreoid and causing the myxœdematous or cretinic degeneration. There are some

features of sporadic cretinism which are not found in the endemic form of the disease. For example, in the endemic form we have found the synthesis of sutures of the bones of the skull. In sporadic cretinism, on the other hand, we find a tendency to lack of synthesis, the fontanelles and sutures remain open a long time, the dentition is delayed, and yet we have the same physiognomy as in the endemic cretin. In the sporadic cretin the skin is pronouncedly myxœdematous, and it will be shown in the author's cases this was one of the chief points in the diagnosis of the disease at an early stage. The endemic type is not pronouncedly myxœdematous. The sporadic form of the disease is manifested by the same tendency to dwarfishness of stature as the endemic form. The anæmia in both forms is similar.

Author's Cases of Sporadic Cretinism.—The cases which have fallen under the care of the writer will be seen to be not only among the youngest cases on record, but they have been observed for a sufficiently long period to enable us to draw some definite conclusions as to the ultimate utility of treatment.

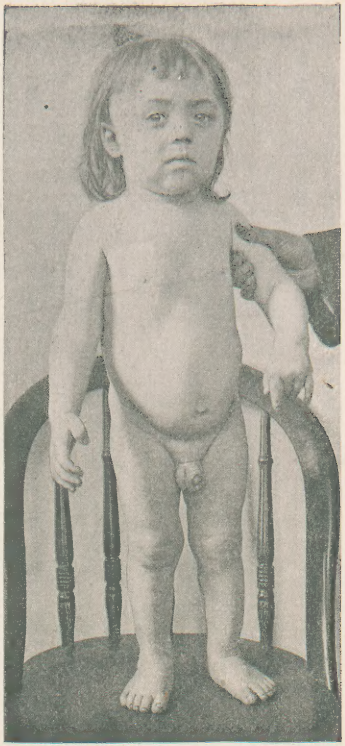
Fletcher Beach, in a discussion before the British Medical Association, presented statistics of fifty-two cases of sporadic cretinism, of which nine cases showed symptoms of the disease in periods from birth to the ninth month; up to the first year, two cases; at the first year, ten cases; up to eighteen months, four cases. Thus almost one half of the cases manifested symptoms of the disease before the eighteenth month.

Case I. *Sporadic Cretinism.*—H. G., male, aged fifteen months, first seen October 10, 1895. Family history shows that the mother and father are first cousins, otherwise they seem healthy; mother is quite a stupid woman,



CASE I.—Before treatment.

After seven months of treatment.



After sixteen months of treatment.

though otherwise does not complain of symptoms of any illness. Mother has no goître, nor has the father. This child was born normally and breast fed; had icterus neonatorum at the age of one week, this lasting four or five weeks. Mother says the child has not at any time been like her other children. He has always seemed stupid for his age, has always been pale, and had a protruding tongue. Has never held the head erect. Has been constipated.

Status Præsens.—Child has an idiotic appearance, a foolish laugh most of the time, is indifferent to surroundings; does not play; has a typical wrinkled, at the same time myxœdematous, skin. The hands are large and flat, saucerlike; the skin is wrinkled over them and cold. The color of the skin is greenish-white; the anæmia is intense; there is an œdematous appearance about the eyes; the bridge of the nose is flattened; the lips are much thickened, and the tongue is thick and protrudes from the lips. The hair is dry, red, and sparse; the forehead narrow; the whole face prognathous; no teeth. There is an inspiratory crow at times, and the voice is a deep guttural. The neck is short and thick; the chest shows nothing abnormal as to contents. Abdomen large, protuberant, and measures fully fifty centimetres. The genitals, scrotum and penis large; the skin of scrotum thickened. Extremities short, as compared to length of body, giving a dwarfed appearance; no deformity.

Internal or rectal temperature, 96°; respiration, 20; pulse, 80.

Hæmoglobin (Fleischl), 18.

White cells, 18,600; red cells, 5,460,000.

After the administration of thyreoid, improvement was noticed within a week. The bowels, hitherto constipated, improved. In two weeks the child was much brighter, the puffiness of the eyelids was reduced; rectal temperature mounted to 97.5°. Hæmoglobin, 30.

In two months the improvement was most marked; the child played with toys; the symptoms of myxœdema about the face were much reduced, and the tongue became less thickened. Hæmoglobin, 45.

After five months of treatment the child was presented at the Academy of Medicine; the child had grown more intelligent; the myxœdema had disappeared; the tongue was still slightly enlarged; the rectal temperature 98° to 98.4°. The hair was replaced by a fine silky growth; the color of the child fair. Hæmoglobin, 55. The teeth have gradually begun to appear. A year of treatment finds an intelligent, chubby child, good-natured, playful; begins to walk along the sofa; has eighteen teeth; tongue is no longer protruded; genitals still large and hands large. Hæmoglobin, 65. Can not talk as yet.

To-day we find a fairly intelligent child, walks and acts like a normal child much younger; does not talk but in monosyllables; is good-natured; no signs of myxœdema; hair long and silky; the tongue is normal; hands still large and flat; the temperature is normal, or almost so. The child is not very strong in its extremities, yet walks unsupported. There is no anæmia that is marked. Hæmoglobin, 85. Received a grain and a half of thyreoid twice daily.

Epicritical Remarks.—In this case the myxœdema, the macroglossia, and the reduction of rectal temperature were marked. The hæmoglobin initial stage was 18, has gradually increased to 75, and the relation of the red to the white cells has increased from 1 in 290 to normal.

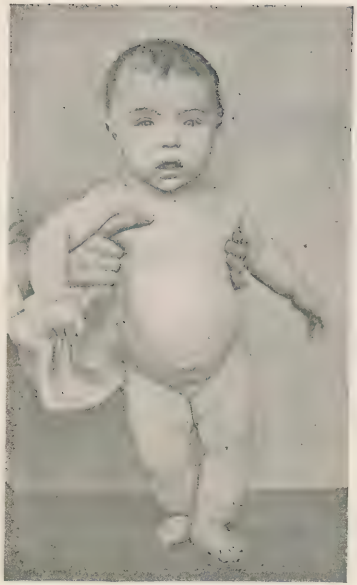
It may be remarked that this child only for the past month has been able to walk about unsupported, this being much delayed as compared with other children. The macroglossia was long on the retrograde, and at first the habit of protruding the tongue persisted, though the tongue from day to day was growing perceptibly smaller. Now, though thirty-one months old and a handsome child, he does not perceive as rapidly as other children the relations of surrounding objects to each other, he smiles intelligently when played with,

and, on the whole, makes a good impression, but not that of a child two years and a half old. He speaks the simple terms—mamma or papa. When he walks it gives us the impression that he is uncertain on his feet. The hair is silky and long, the lips are no longer thick, nor is the tongue; the abdomen is not protuberant, and the relative length of the abdomen and extremities seems proportionate. The hands alone still seem a little large and flat, and the child is easily chilled, especially the extremities.

It should be noted in the history that the child was jaundiced after birth, and it will be seen that another case, his younger sister, presented symptoms of cretinism at birth, was also jaundiced at this time, as also the following case:

Case II. *Sporadic Cretinism.*—Symptoms of myxœdema especially marked. Female child, aged twenty months, first seen April 10, 1895. Family history shows that the mother and father are cousins, one remove. Mother and father healthy, no goître; but the mother has a slight, scarcely noticeable fullness over the isthmus of the thyreoid; this appeared during her pregnancy with this infant. Infant is breast fed; when two weeks old was jaundiced; this continued three or four weeks. When three months old had adenitis of glands in the neck. At the age of six months the mother says the infant became more and more stupid and weaker physically. It had attempted to walk when a year old, but now it made no effort to do so. During the last two months the child had developed abscesses on the hands and feet which refused to heal. The face of the child had changed so markedly as to be noticed by strangers.

Status Præsens.—Mentally a very stupid child, takes notice of nothing about it; attention can not be drawn to bright objects or striking colors.



CASE II.—Before treatment, aged twenty months.

After a year of treatment.

After twenty-two months of treatment.

Skin has a greenish-yellow, waxy appearance, especially in the face; can not hold its head erect for more than a moment; hair sparse and very dry. The upper and lower eyelids are much swollen, and bridge of the nose flattened; puffiness of the cheeks, and the lips are thickened. Decided thickening of the tongue; voice guttural; no teeth. Abdomen protuberant, and extremities small and dwarfed; skin thickened, but not truly œdematous, more myxœdematous. Chest organs negative; circulation very sluggish, as the skin about the abscesses to be described is bluish in tint. Rectal temperature, 96°; pulse, 100; respiration, 22; hands and feet cold; body cool; bowels constipated.

Extremities: Skin over extremities finely wrinkled and myxœdematous.

Abscesses: On the index finger of the right hand and the lower third of the left forearm, on the left thumb, and the dorsum of the left foot were abscesses which simulated syphilitic bone lesions (dactylitis). There were small abscesses with bluish discolored bases, discharging a creamy white material. It developed that they had been treated for months without effect. Urine negative. Hæmoglobin, 25 (Fleischl).

Thyreoid treatment improved the child within two weeks. The anæmia, myxœdema, and abscesses seemed to improve in this short time; the rectal temperature rose a degree. After a month the child was brighter and the abscesses on the extremities had healed; the myxœdema had almost gone. Hæmoglobin, 40. Child tries to sit up, and two months after beginning of treatment cut its first tooth, and tried to sit up in its chair.

July 31st, or three months after beginning of treatment, child had an attack of gastro-enteritis. The thyreoid suspended, and with this the infant did not have therapy of the thyreoids for two months. The symptoms of myxœdema gradually returned, the stupidity, puffiness of the face, thickening of the lips and eyelids, constipation, and lastly the abscesses reopened. The thyreoids begun again, and improvement was immediate and steady, and has continued up to the present day.

October, 1896.—Rectal temperature, 98.4°. Hæmoglobin, 70. We find a bright child walking; no anæmia that is marked; no symptoms of myxœdema; began to walk two months ago. The child is very intelligent, plays, and tries to talk; the talk is not perfect, but she can make herself understood. We still see a child fully three years and two months old impress us as a normal child of two years. Thyreoids at present, two grains a day.

March 1, 1897.—Hæmoglobin, 85.

Epicritical Remarks.—In this case we have a predominance of the myxœdematous symptoms—that is, the œdema and puffiness of the face, skin, body, and extremities seemed to be of the typical character known in the adult as myxœdema. The child was in good health until the sixteenth or seventeenth month, when it developed abscesses over different portions of the body, when it came under observation, as will be seen by referring to the history; some of these abscesses were on the fingers and back of the hand, resembling those seen in congenital syphilis. They differed from them in that the skin around the summit of the abscess was of a dark bluish tint, due to the sluggish circulation in the presence of a reduced body temperature. The discharge from the abscess could not be called pus; there was a whitish creamy fluid, which, when examined under the microscope, showed detritus and a few epithelioid nuclei. They healed under the thyreoids, and they in every way answered to the description of similar foci of necrosis found in subjects of endemic cretinism. A most interesting part of the history in this case was the return of all the symptoms of myxœdema after a temporary suspension of the treatment. The anæmia, the myxœdema of the face and extremities, the reduced body temperature, and even the

abscesses, these improved again when treatment was resumed. Though the child walks and talks to-day, it is far behind a child of its own age (three years and a half) in normal state. Though intelligent, it is not very bright; it has an intelligent smile and laugh when played with. It walks without support, and talks sentences of several terms. The voice has a normal timbre, but the child is fully one year behind other children in development, and one looking at the child would take it for a normal child about two years and a half old. Thus in psychic development it has lost as much as it was advanced in the disease when it came under treatment. This child is also easily chilled. Its hands at times seem much colder than those of normal children under like conditions of weather. It notices more the relations of surrounding objects and persons than Case I, and is not in any way hard of hearing. The skin and hair are normal.

Case III. *Congenital Sporadic Cretinism or Myxœdema.*—Female infant, aged one month, seen first on December 24, 1896. This infant is a sister to the patient in Case I, and is the first infant born to the mother after the cretin, Case I.

The birth was a normal one. When four days old, jaundice neonatorum appeared and persisted six weeks. At the age of a month the mother brought the infant to me for the jaundice.

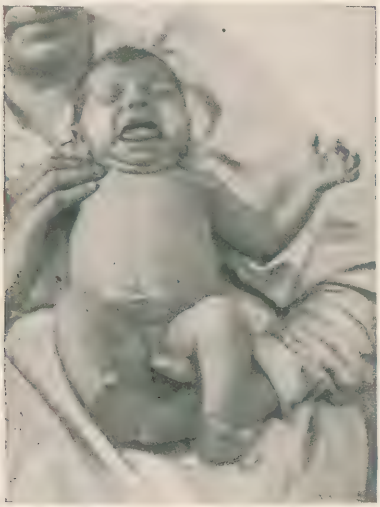
I saw a fairly nourished infant still jaundiced. The expression of the face was striking; the infant did not cry unless severely teased; it seemed very torpid. The head was round, broad at the base of the skull, smaller at the summit, not markedly pointed. The abdomen was distinctly rotund; the extremities were short, but not deformed. Child was short and thickset. There was no myxœdema of the skin, but the whole surface was cool to the touch; lips were slightly puffed; tongue was very large and thick—macroglossia; neck short and thick.

Head: Circumference, 38 ctm.; antero-posteriorly, 23 ctm.; bitemporal, 20 ctm.—rather of the oxycephalic type.

Fontanelle ant., $3\frac{1}{2} \times 2$ ctm. Sagittal suture open to the occipital depression.

Thyreoid not palpable.

Hæmoglobin, 85. Temperature, rectum, 96°.



Case III.—Congenital sporadic cretinism in a child a month old.

As said above, child is stupid, and clapping of the hands failed to attract its attention. Under thyreoid treatment the temperature gradually rose in the rectum to 98°. The infant became bright and seemed to notice surroundings and smile. The tongue is becoming markedly thinner, and the face is taking on a normal expression. It plays and laughs and cries as other infants; forehead is broader and not wrinkled.

February 15th.—Hæmoglobin, 60. Temperature, 98.4°, rectal. In this case at no time could thyreoid be felt and no supraclavicular masses of fat were present.

In this case we find a newborn babe, whose mother had previously given birth to a cretin, showing symptoms of slight cretinism. The stupidity of the infant, the reduced internal temperature, the peculiar conformity of the extremities as related to the trunk, the thickened tongue, the wrinkled skin on the forehead and hands, the short animal forehead, the thick lips, the immense tongue (macroglossia), coarse cry, the immediate improvement under thyreoid of the stupidity and the thickened and hypertrophied tongue, all point to the inevitable conclusion that here we have to deal with a congenital cretin, who, if allowed to progress, would develop the full symptoms of the disease.

It is an interesting fact that in this case, early in the disease, the hæmoglobin was greater than later on, though the infant was immediately placed upon thyreoids. This would seem to point to the fact that the anæmia of cretinism develops as the disease progresses, and is not present at the initial stage of the disease.

Thus in all the cases brought forward by the writer in this paper as sporadic cretinism the disease was diagnosticated at a very early period, and in the third case at a period immediately after birth before the disease had time to fully develop. They have been under observation, two of them, for a period sufficient to demonstrate that though the thyreoid treatment rescues these unfortunates from a state of perpetual idiocy, it does not restore fully the psychical state which has become dwarfed for a greater or less period before the therapy was initiated. Though bright, the children are not the equals of children of normal condition of their own age, but are very slow in appropriating ideas and in perfecting their speech vocabulary. They seem prone to attract to them-

selves the least ailments; both of our cases, I and II, having contracted diphtheria just as other children, and have recovered. We have seen in all the cases the thyreoid gland could not be made out. The masses of supraclavicular fat were not present in any of our cases. There were in the first two cases very pronounced symptoms of myxœdema; in the newborn infant this was not the case. The stunted growth was marked in all the cases, and in Case II the photograph shows the marked disproportion of the lower extremities and length of body. In all cases the therapy repaired this defect. The voice in all three cases was of a barking, hoarse nature, and in two of the cases this voice persists. The hair was coarse and dry in two of the cases, and was replaced under therapy by a silky growth.

Anæmia was marked in the two advanced cases, but a blood count failed to establish a marked leucocytosis, which Horsley and Kocher found in the myxœdema produced by extirpation of the thyreoid in the monkey and human subject. The marked improvement of the anæmia was pronounced in the advanced cases, and in the newborn infant was not immediate—on the contrary, seemed to progress until the correct dosage of thyreoid was found.

The psychical status in all cases was stupid and reduced to the idiotic; even the newborn babe would only react when irritated, and then would relapse into a stupid sleepy condition.

The rectal temperature was reduced in all cases, and immediately rose when the thyreoids were given. The reduction of temperature was full two degrees below the normal in all cases.

Cases which simulate but are not true cretinism have

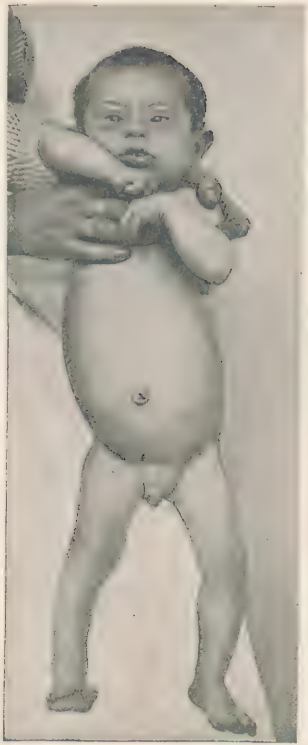
been recorded from time to time by observers as either true cretinism or pseudo-cretinism. There is an objection to the term pseudo-cretinism, but there are forms of disease which, if not true cretinism, are still very difficult to classify.

In this paper I shall consider the following forms of disease, and try to point out in what way they differ from the true cretinoid degeneration:

1. The dwarf with idiocy.
2. Mongolian idiocy.
3. Lipomatosis universalis (a form of partial idiocy and fat accumulation).
4. The peculiar state of hydræmic anæmia in children as distinguished from myxœdematous degeneration.

The Dwarf with Idiocy, or Partial Arrest of Intellectual Growth.—A case of this kind we found in our clinic. The child was ten years and a half of age, born in a foreign country of healthy parents. Father's sister has epilepsy; the other children healthy. We find in this case a perfectly well-formed individual, with well-proportioned hands and feet. The head is not badly shaped, though tending to the brachycephalic type. The child has a slight anæmia, and this is probably due to lack of proper diet. There was not the remotest indication of myxœdema. The dwarf is intelligent, wishes to learn, and plays with other children. She has only talked for the past year, and now talks short sentences well. The internal temperature is normal. The child was brought to the clinic for nosebleed. It seemed to the author that the retarded intellectual condition was due as much to the neglect of the parents, who looked upon the little one with disfavor, as to the dwarfed brain. The head

Dwarf with retarded mental development, aged ten years and six months.



A Mongolian idiot aged five months.

Lipomatosis universalis, child aged ten years.

was large if anything. The child was strong, and the only fault it possessed was that when irritated it would fly into a passion. The expression of the face is not cretinoid and is not prognathous. Thus we have none of the symptoms of cretinism, and yet the case was brought to the author as one of probable cretinism. We find in it only an exquisite example of the dwarf with weak intellectual status.

The Mongolian type of idiocy has probably been more than any other form of disease improperly classed with the cretins. At a recent meeting of the British Medical Association Tedford Smith and Beach presented papers in which the Mongolian idiot was fully described and the effect of thyreoid treatment discussed. The idiots of the Mongolian type resemble the cretins quite closely. There is the stunted growth, the open mouth, thick lips, and large tongue; in some the tongue is being constantly protruded; the hoarse guttural voice, mental apathy and sluggishness; sometimes subnormal temperature; in others I have found the temperature normal. The skin is dry and hair coarse, but in the young infant not particularly so. The musculature is quite flabby; the occipital part of the skull quite deficient, the neck short and thick. The infants of this type are so weak as to be unable to hold their heads erect. Frequently there is strabismus, and the eyes have the distinctly Mongolian type. The head is small and mostly brachycephalic, as can be seen from the portrait; the fontanelles remain open long, and dentition is delayed. But there is no myxœdema, and the hands, though flat, are not saucerlike, as those of the cretin; the small finger of both hands is bent slightly at an angle internally. The anæmia may or may not be as pronounced as that of the cretin, but there is not that

greenish hue to the skin, nor is the prognathous face as marked. Though the muscles are flabby, there is no deformity of limb. Their improvement under thyreoid is still a matter of discussion.

Idiocy of the Mongolian Type with Macroglossia.—Male infant, aged five months, first seen May, 1896. There is nothing of note in the family history. The labor was rapid, infant small; brought up on artificial means.

The infant has an idiotic expression to its face, there is convergent strabismus, and the tongue protrudes, showing the macroglossia. The infant does not hold its head upright, but nods it from side to side. The head is broadest in its biparietal diameter, and flat on top and also flat at the occiput. The fontanelles are all open and the sutures quite wide. The neck is short and thick, and this can best be seen from behind. The skin is not myxœdematous or cool. The hair is sparse and dry; no teeth; abdomen quite large; extremities long and thin, not of the cretin type; musculature very soft and flaccid. Infant notices, but in a semistupid manner. Eyes converge, giving the Mongolian type to features. Rectal temperature, 99.8°; hæmoglobin, 25.

The above brief *résumé* will serve to show how distinct from cretins this type is, yet hardly characteristic of the form of degeneration. When these infants grow up they maintain the close resemblance to the portrait, and all of these idiots seem to have a similar appearance.

Lipomatosis Universalis.—The following case is an example of an exceedingly rare condition of childhood. This case was originally in the service of Dr. Scharlau, in the Mount Sinai Hospital, and was referred to my dispensary for outdoor treatment. The patient was under

my care for over a year, and then passed to another hospital and was published in *Pædiatrics* of 1896, by Dr. Dessau. The case was recognized by the writer as one of lipomatosis universalis, and so treated. In many respects it is similar to cretinism. The head of the patient is small, the forehead low, but here the resemblance ceases. There is no prognathic face, and there is no myxœdema, the skin being warm and smooth, and there is no anæmia. As seen, the accumulation of fat is enormous. The intellect is that of a quasi imbecile, yet the boy talks quite intelligently on simple subjects. The muscular tremor and weakness, as also partial blindness, seem more a result of lipomatose degeneration.

Lipomatosis Universalis, with Blindness and Muscular Weakness and Mental Arrest of Development.—Boy, aged ten years, first seen February 18, 1895. Family history, on the whole, good; no neurotic tendencies. Birth of this patient normal; breast fed; healthy infant, not markedly fat. Walked at the age of one year, and talked when two years of age. Always sleepy, and in this respect differed from other children. At the age of six years sustained a fall on the side of the head. About this time began to grow so stout, especially about the abdomen, as to cause the mother to go to a physician for advice. His eyesight next began to fail, and he finally has become quite blind. Tremor of hands and weakness have also appeared with great increase of fat.

Status Præsens.—We find a boy of ten years with a fat accumulation reminding one of an obese adult. The head is small, with a narrow, retreating forehead and small occiput. The hair is dry and curling, the face is flat and broad; the expression worried, as if he did not see, dull, stupid in repose; ears large; sense of taste and touch very highly developed; hearing acute; speech is deep-voiced; tongue normal. There are tremors in hands and lower extremities on volition, as in button-

ing his coat, but not in repose. Reflexes at patella and ankle exaggerated. Mentally bright when talked to, stupid if left alone. If left alone, will sleep constantly, and will only rouse when touched; will sleep for fully twenty-four hours.

Physically the fat accumulation is enormous; weighs a hundred and sixteen pounds. Initial weight, ninety-nine pounds when first seen.

Height, 129 ctm.; head, glabella to occiput, 53½ ctm.; breast, 81 ctm; arm, 28 ctm.; forearm, 21 ctm.; wrist, 16 ctm.; abdomen, 95 ctm.; thigh, 49 ctm.; leg, 53 ctm.; ankle, 24 ctm.

Skin over body is dry; hair dry, grows very slowly, and nails grow very little; never pares the nails.

Mentally: Fears the approach of strangers, as he is blind and doesn't know whether they are friendly; when assured, is affectionate. Memory is very good; is intelligent; can count change by touch.

Gait: Is that of a very fat individual.

Eyes: Examined by Dr. William Cowan. Marked atrophy of the optic nerve and retinæ; opacities of the vitreous.

Heart: Weak, and gives impression that both sounds are alike; the muscular part of first sound not present. Irregularity in rhythm.

Genitals: Atrophied. Urine: Nothing abnormal. Appetite ravenous. Thyreoid treatment; no effect.

The above are only excerpts of a very lengthy history.

The last series of cases which I desire to describe are a peculiar class of hydræmic anæmias with a peculiar swollen condition of the eyelids, face, and lips, sometimes of the extremities, which might by the inexperienced be taken for a beginning myxœdematous degeneration. Fortunately, the hydræmia in the special set of anæmias here recorded rarely extends to other parts of the body. The internal temperature is normal, and these subjects are exceedingly bright intellectually. In neither of the

recorded cases was albumin found in the urine. In short, there is only a facial semblance to the myxœdematous degeneration. The bony system is perfect in both cases; even in the face we find no obliteration of the bridge of the nose; no macroglossia; no affection of the hair, nor is the skin harsh or dry.

We append two cases of this affection with portraits and brief histories:

Case I. *Hydræmic Anæmia.*—Male infant, aged fifteen months; seen first October, 1896. Family history negative. Infant is breast fed; is now beginning to walk. He is a fairly bright infant, notices objects and plays.

Hydræmic anæmia, child aged fifteen months.

Is exceedingly anæmic; skin is of a greenish-white hue; there is not only puffiness under the eyelids, but the lips are swollen, and when at rest the face has a stupid ex-

pression. The extremities are well proportioned as compared to the body, and there is a mild form of rhachitis. Abdomen large; spleen slightly enlarged. Rectal temperature, 98.2°; urine negative. Hæmoglobin, 30.

Case II. *Hydræmic Anæmia.*—Female child, aged eight years, first seen May, 1896. Birth was normal; infant breast fed; has always been pale and weakly. Is now a fairly well-developed girl. Has an extreme form

Hydræmic anæmia, child aged ten years.

of anæmia; skin of a claylike color; there is swelling under the eyelids; puffiness of the face and lips, not of the extremities. Hair glossy, not dry. Angle of eyes give the features a Mongolian type. Mucous membranes very pale. Musculature badly developed. Signs of early rhachitis, otherwise body well formed. Mentally, a very bright child, and satisfactory at school. Hæmoglobin, 55. Urine negative. Red and white cells, 483 to 1. Temperature normal.

From the short excerpts given of the histories of these two interesting cases it will be seen that they resemble a myxœdema only in the puffiness about the skin of the

face as detailed, but here the resemblance stops. There are none of the characteristics mentally, or in the skin of the rest of the body, or in the bones, to bring these cases in the same class as the cretinoid degeneration.

The œdemas due to nephritic disease are not so apt in children or infants to be mistaken for a myxœdematous condition.

An examination of the urine in infants and children will eventually clear up the case. In adults, M. A. Starr has related cases where myxœdema was mistaken for Bright's, or even cases where albuminuria with myxœdema were coexistent, yet in children the kidney disease presents no difficulties of diagnosis. Nor are we likely to have, as in the adult, marked kidney changes without positive evidences in the urine; such cases must be exceedingly rare.

In conclusion, the author wishes to express his obligations to Dr. Emily Lewi for untiring industry in following up the history and therapy of these cases and making all the blood examinations at various intervals.

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